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## CASE REPORT

# Adventitial cystic disease of the common femoral vein presenting as deep vein thrombosis



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## KEYWORDS

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**Summary** Adventitial cystic disease of the common femoral vein is a rare condition. We herein report the case of a 50-year-old woman who presented with painless swelling in her left lower leg that resembled deep vein thrombosis. She underwent femoral exploration and excision of the cystic wall. The presentation, investigation, treatment, and pathology of this condition are discussed with a literature review.

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## 1. Introduction

Adventitial cystic disease (ACD) of the veins is a rare condition with an uncertain etiology in which a mucin-containing cyst is formed in the walls of the veins. The disease may be difficult to diagnose because its incidence is

very low and the initial presentation is similar to that of deep vein thrombosis (DVT).<sup>1–3</sup> We herein report the case of ACD of the common femoral vein (CFV), which showed characteristic features similar to that of DVT, in a 50-year-old woman who presented with painless swelling in her lower leg. This case report was approved by the investigational review board (Research No. KC11ZISE0635).

## 2. Case report

A 50-year-old woman presented with a 1-month history of unexplained edema in the left lower leg. The circumference

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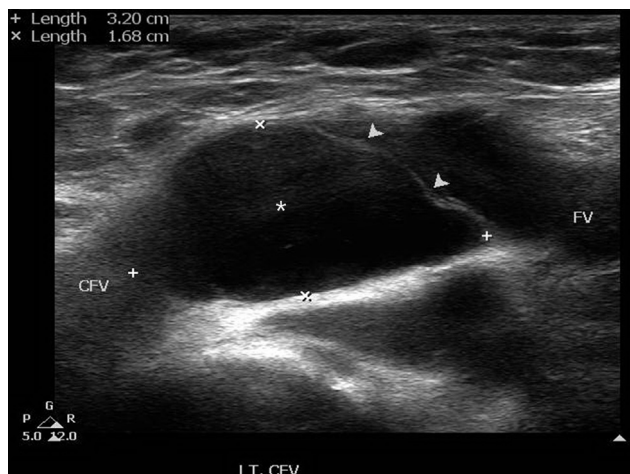
of the right thigh was 3 cm larger than that of the contralateral side at the mid-thigh level. On the day of admission, her serum D-dimer level was normal (0.26 mcg/mL), but ultrasound examination revealed the presence of an anechoic focal mass, measuring approximately 3.2 cm  $\times$  1.7 cm, in the left CFV without compressibility (Fig. 1). A contrast-enhanced computed tomography (CT) scan also showed the presence of an intraluminal low-attenuating mass lesion (2.7 cm  $\times$  1.8 cm) involving the left CFV (Fig. 2).

In view of presumed DVT, the patient was anticoagulated with low-molecular-weight heparin; a retrievable inferior vena cava (IVC) filter was placed at the infrarenal segment by the right internal jugular venous approach, after which surgery was done for exploration. While performing the surgery, the common femoral artery was exposed and retracted laterally to facilitate dissection of the CFV. The CFV, deep femoral vein, and great saphenous vein (GSV) were dissected. When a transverse venotomy was performed on the CFV, a 3.0 cm  $\times$  1.7 cm adventitial cystic mass extending from the medial to the posterior surface of the left CFV and the vein lumen was found in a compressed state (Fig. 3). After the gelatinous mucoid substance was evacuated from the cyst, the lumen of the CFV showed no persistent stenosis and pathologic scarring. We resected the cyst wall, except the parts attached to the CFV, with a simple closure of the venotomy site in a transverse fashion using GOR-TEX CV number 7 suture (W. L. Gore & Associates, Inc., Flagstaff, AZ, USA). Pathological results identified the mass as an adventitial cyst.

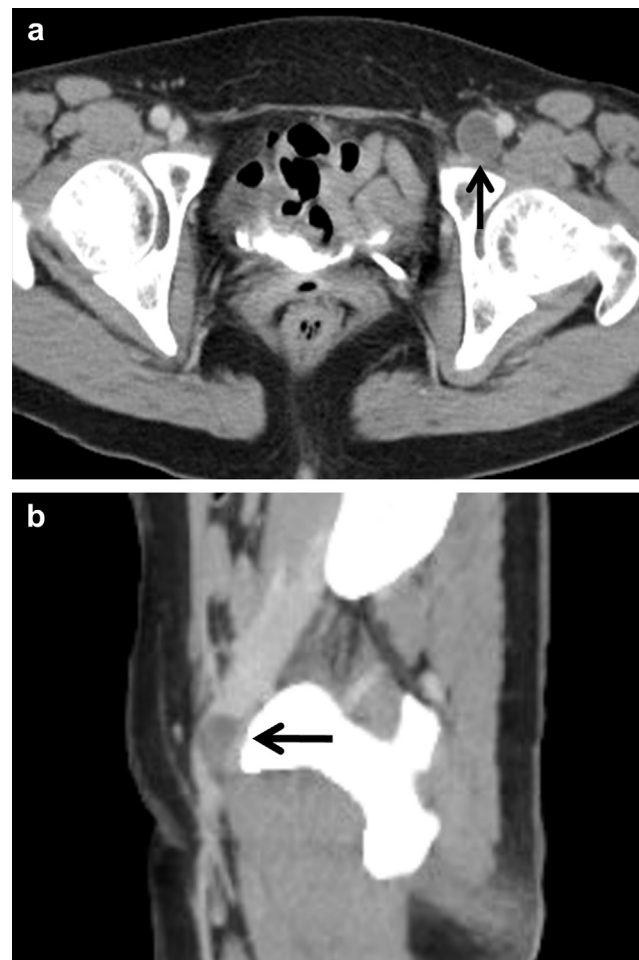
An improvement was observed in the edema of the patient's leg after surgery and the IVC filter was retrieved percutaneously the next day. The patient was discharged on postoperative Day 2 without being prescribed any anticoagulants.

### 3. Discussion

ACD is characterized by the accumulation of a gelatinous fluid containing mucoproteins and mucopolysaccharides



**Figure 1** Ultrasound longitudinal scan showing the presence of a 3.2 cm  $\times$  1.7 cm, noncompressible, anechoic mass lesion (asterisk) involving the left common femoral vein, and compressing the venous lumen.

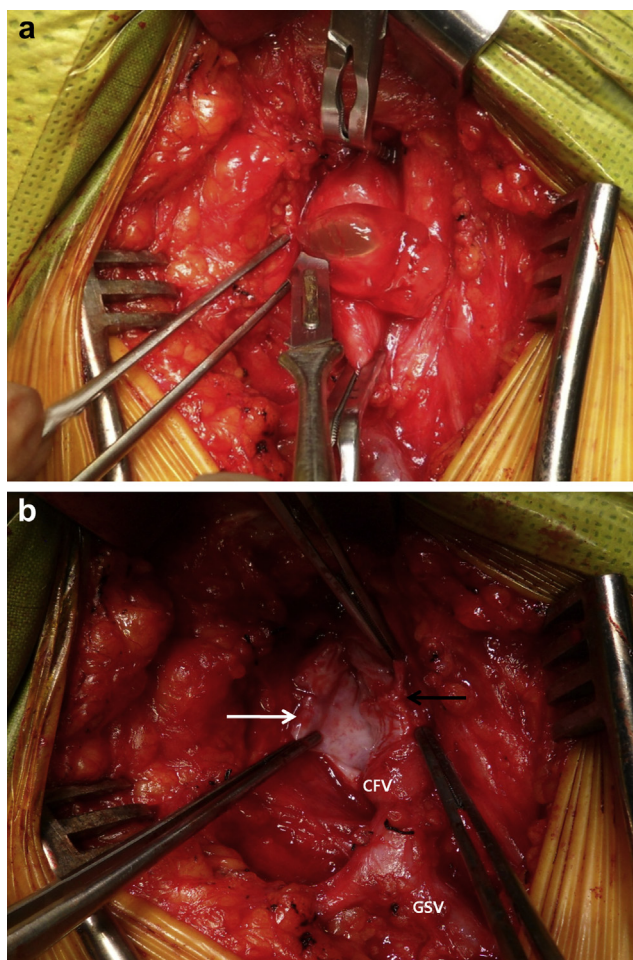


**Figure 2** Contrast-enhanced computed tomography demonstrating distension of the left common femoral vein due to an intraluminal hypoattenuating mass lesion (arrow) attached to the posterior wall. The mass was presumed to be a deep vein thrombus.

within the adventitial layer of the blood vessel.<sup>4</sup> ACD of the venous system is a very rare condition, with only about 27 cases described in the worldwide literature. Maldonado-Fernández et al<sup>5</sup> summarized 18 cases of venous ACD in 2004, which were all reported prior to 2001. In this report, we summarized an additional nine cases, which were reported after 2001 (Table 1).

The exact etiology and pathogenesis of venous ACD still remain uncertain. However, it can be explained in similar terms as that of arterial ACD: (1) the developmental theory (mesenchymal cells from nearby joints implant into the adventitia of the vessel during embryological development); (2) the repeated trauma theory (the adventitia undergoes cystic degeneration as a result of stretching and distortion near the joints); (3) the systemic disorder theory (degeneration of the adventitia as a result of connective tissue diseases); and (4) the ganglion theory (synovial cells implant into the adventitia near the joints).<sup>4,6</sup>

According to the review of 27 cases (Table 1), ACD of the vein tends to develop at a later stage in life (23–75 years; mean: 46.7 years). Among the 27 patients, 16 were men and



**Figure 3** (a) Operative image illustrating the gushing of gelatinous mucoid content from the incision of the CFV. (b) An adventitial cystic mass (white arrow) extends from the medial to the posterior surface of the left CFV. The black arrow indicates a simple closure of the venotomy site in a transverse fashion with CV number 7-0. CFV = common femoral vein; GSV = great saphenous vein.

11 were women (M:F ratio: 1.5:1). The most commonly affected veins were the femoral vein (51.9%), external iliac vein (18.5%), CFV and external iliac vein (7.4%), popliteal vein (7.4%), small saphenous vein (7.4%), GSV (3.7%), and wrist (3.7%) veins. Macroscopically, cysts are either uniloculated (70.4%) or multiloculated (29.6%) and filled with clear or yellow mucoid material between the media and adventitia. Microscopically, cysts may or may not have an epithelial lining. As fluid accumulates within the cyst, it compresses the vein lumen, resulting in stenosis or occlusion.

Because the most frequent symptom of venous ACD is swelling of the affected limb, it is very difficult to differentiate venous ACD from DVT. Therefore, ACD of the vein should be considered in the differential diagnosis of lower extremity edema.

There is no optimal investigation for the diagnosis of venous ACD, but several imaging modalities have been reported to be useful. Ultrasound is typically the first-line imaging modality in the assessment of thrombosis in deep

venous structures.<sup>7</sup> Ultrasound imaging may show the presence of a typical, anechoic mass with a posterior acoustic window and may allow ultrasound-guided treatment.<sup>8,9</sup> By contrast, ultrasound imaging in DVT may reveal the presence of a hypoechoic or hyperechoic, thrombus-filled vein with obstructive flow. However, the presence of ACD and that of isolated DVT are not mutually exclusive, as in our case.

A CT venography can be successfully used in imaging ACD of the vein. When compared with venography, it has the advantage of a noninvasive examination that can directly image the surrounding parenchyma and aid in surgical or percutaneous treatment planning. A magnetic resonance imaging can reveal the presence of high-signal-intensity cysts with extrinsic compression of the vessel lumen.

There are three options for venous ACD treatment. First, minimally invasive management has been reported with image-guided drainage of adventitial cysts, but incomplete evacuation of cysts secondary to high viscosity has resulted in high recurrence rates. When deep veins were involved, puncture and drainage were done in three cases, but were followed by recurrence in two patients.<sup>5</sup> However, Johnson et al<sup>9</sup> reported a case of successful treatment of ACD of the femoral vein without any recurrence by percutaneous aspiration and sclerosis. Therefore, percutaneous aspiration and sclerosis may be recommended to patients with high risks of surgical procedures. Second, the preferred surgical management of venous ACD is either transadventitial or transluminal evacuation of the mucoid cyst with excision of the cystic wall. These are excellent options when there is no associated venous thrombosis, wall thickening, or persistent venous stenosis after drainage. However, recurrence may be an issue in cases of incomplete enucleation.<sup>4</sup> In our patient, the CFV was patent, and therefore, the ACD wall was segmentally resected with a simple repair of the venotomy site. Consequently, close follow-up of the patient will be necessary to ensure a successful outcome. Third, the most definitive treatment is a complete resection of the cyst and the involved vein, followed by venous reconstruction, preferably with vein patch, interposition vein, or polytetrafluoroethylene graft, and anticoagulation treatment. However, no particular regimen has demonstrated superior results according to the limited follow-up data of past case reports.

In general, optional vena cava filters can be considered in certain indications, such as ilio caval DVT, large free-floating proximal DVT, massive pulmonary embolism (PE) treated with thrombolysis, or thrombectomy.<sup>10,11</sup> In this case, optional vena cava filter was not indicated. However, we inserted temporary vena cava filter because we thought PE could develop during CFV exploration or surgical thrombectomy. After the operation, the IVC filter was retrieved.

In summary, we reported a case of successful transluminal evacuation of a mucoid cyst and excision of the cystic wall of ACD from the CFV, which was initially diagnosed as DVT. Thus, ACD of the vein needs to be considered in the differential diagnosis of unexplained leg swelling. Furthermore, to ensure a successful outcome, close follow-up of the patient is necessary.



**Table 1** Cases of cystic adventitial disease in veins documented after 2004.

Study reference	Gender, age (y)	Location Thrombus in vein	Nature	Treatment	Recurrence (follow-up duration)
Gasparis et al <sup>3</sup>	M, 37	Left external iliac vein Thrombus (+)	Multiple cysts	Excision of vein, anticoagulation (+)	No
Sugimoto et al <sup>2</sup>	F, 48	Right CFV Thrombus (–)	Single cyst	Excision of vein, anticoagulation (+)	No (1 y)
Sakamoto et al <sup>12</sup>	F, 56	Right popliteal vein Thrombus (–)	Single cyst	Excision of cyst wall, anticoagulation (–)	No (4 y)
Dix et al <sup>8</sup>	M, 28	Right CFV Thrombus (+)	Single cyst	Excision of cyst wall, anticoagulation (+)	No (6 mo)
Seo et al <sup>13</sup>	M, 69	Left CFV Thrombus (–)	Single cyst	Excision of vein, anticoagulation (–)	No
Johnson et al <sup>9</sup>	M, 65	Right CFV Thrombus (–)	Single cyst	Aspiration only, percutaneous aspiration and sclerosis, anticoagulation (–)	Recurrence (after 6 mo) No (6 mo)
Morizumi et al <sup>14</sup>	M, 28	Left CFV Thrombus (–)	Single cyst	Excision of cyst wall, anticoagulation (–)	No (6 mo)
Jayaraj et al <sup>15</sup>	M, 36	Left CFV Thrombus (–)	Single cyst	Excision of vein, anticoagulation (–)	No (6 mo)
Present case	F, 50	Left CFV Thrombus (–)	Single cyst	Excision of cyst wall, anticoagulation (–)	No (3 mo)

CFV = common femoral vein; F = female; M = male; mo = months; Y = years.

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